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Case Report

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Recurrent cardiac papillary fibroelastoma with multiple organ embolism. Is it really benign?: a case report

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ABSTRACT

Cardiac papillary fibroelastomas are the second most common benign heart tumor after myxomas. They are usually asymptomatic but may cause embolic symptoms. In this case report, we present a patient who developed acute thrombosis in the lower extremity and was operated for cardiac papillary fibroelastoma in addition to embolectomy. In addition, tumor recurrence developed 2 years later, multiple embolisms occurred in the kidney and spleen, and aortic valve replacement was performed.

Keywords: Cardiac papillary fibroelastoma, heart tumor, embolism

INTRODUCTION

Cardiac papillary fibroelastomas (CFE), accounting for <10% of all cardiac tumors, are the second most common benign heart tumor after myxomas.[1] They are usually asymptomatic and diagnosed incidentally. However, according to the localization and size of the tumor; stroke, myocardial infarction, pulmonary embolism, embolism in extremity arteries, renal artery and splenic artery may occur, sudden cardiac death may develop.[2] In this case report, we present a patient who first developed acute thrombosis in the iliac artery, therefore embolectomy was performed and then cardiac mass was removed, and two years later, kidney and spleen infarction developed and aortic valve replacement (AVR) was performed due to the recurrence of the tumor in the aortic valve.

CASE

Patient consent was obtained for this study. A 50-year-old female patient was admitted to an external center with the complaint of pain in her right leg and embolectomy was performed due to the detection of acute thrombus in the right iliac artery. She was referred to us after a 12x12 mm mass was detected at the level of the aortic valve in the transthoracic echocardiography (TTE). Her vital signs were stable, and there was no abnormality in her examination, history and laboratory parameters. However, due to sudden onset of pain, coldness and color change in the left leg, left femoral embolectomy was performed. She was then operated with a median sternotomy, and a fragile, lobulated mass extending over the aortic valve noncoronary leaflet was removed. No dysfunction was observed in the aortic valve. Fibrous material was detected in the pathology report of the operation material. After the left femoral embolectomy, the material sent to pathology was also found to be fibrinous material. The patient did not develop any complications in the follow-up, and was discharged with 300 mg of acetylsalicylic acid (ASA).

Two years later, a solid mass of 10x9 mm was detected in the left leaflet of the aortic valve in the TTE of the patient who developed dyspnea. Ejection fraction: 40%, mild mitral regurgitation, moderate aortic regurgitation, and a maximum gradient of 27 mmHg in the aortic valve were detected. Thereupon, cardiac magnetic resonance (MR) was performed to evaluate the recurrence of the mass and other cardiac cavities, and a solid mass was observed in the same place (**Figure 1**). In addition, bilateral renal hypoechoic areas were detected in abdominal ultrasonography. On computed tomography of the abdomen, there was an ischemic lesion area of embolism and infarct areas in the right kidney in the middle and upper pole posterolaterally. There were ischemic areas in the upper pole posterior and lower pole of the left kidney, but no infarction had developed yet (**Figure 2**). In addition, an infarct area due to embolism was detected in the posterosuperior part of the spleen (**Figure 3**).

Thereupon, it was decided to re-operate the patient. Median sternotomy was performed at the same place, and there was a 1x1x0.5 cm rough, hard mass on the right aortic leaflet with a broad base, extending between the right and left leaflets, and restricting the movement of both leaflets. The mass

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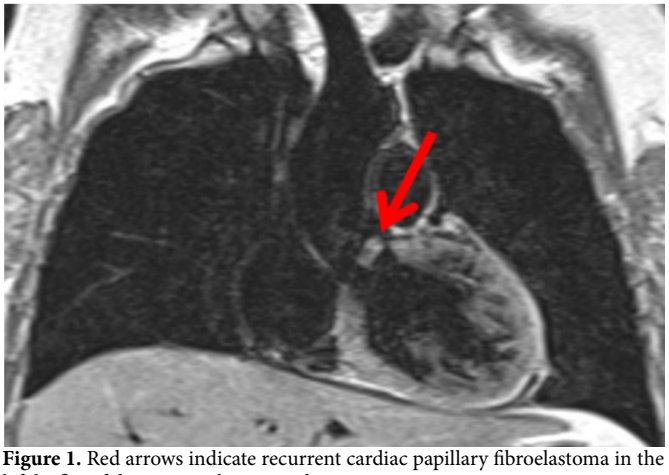

Figure 1. Red arrows indicate recurrent cardiac papillary fibroelastoma in the left leaflet of the aortic valve on cardiac magnetic resonance

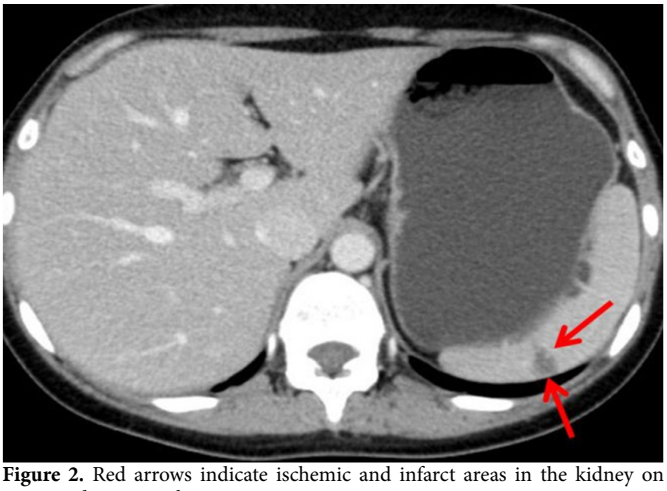

Figure 2. Red arrows indicate ischemic and infarct areas in the kidney on computed tomography

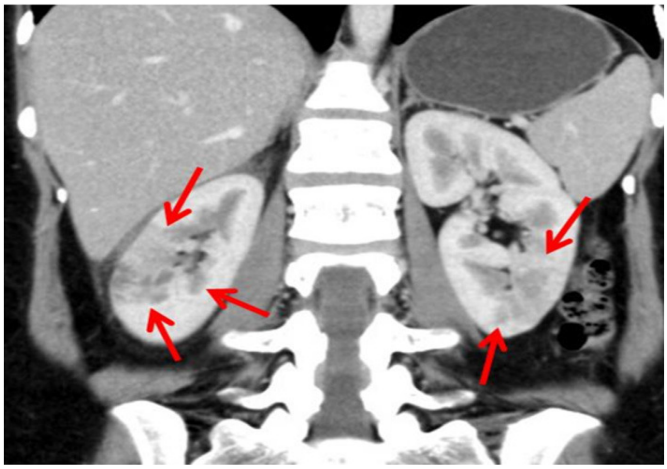

Figure 3. Red arrows indicate the area of infarct due to embolism in the posterosuperior part of the spleen on computed tomography

was excised together with the dysfunctional aortic valve. The aortic root was enlarged with the Nick technique using a synthetic graft and a number 23 St. Jude mechanical valve (St. Jude Medical, Minneapolis, USA) was inserted. In the pathology report, it was observed that fibrous connective tissue was formed in the blood-fibrin association. In the follow-up, the patient was extubated on time and discharged with ASA 300 mg and warfarin.

DISCUSSION

Primary tumors of the heart are less common than secondary tumors. CFE comes after myxomas, which is the most common heart tumor, and is the second most common heart tumor and has a benign character.[1] CFE consists of a small papillary, pedunculated, avascular tumor covered by a single endothelial layer containing variable amounts of elastic fibrils in the form of collagenous connective tissue in the hyaline stroma.[3] The worldwide prevalence of CFE during autopsies and open heart surgery ranges from 0.02% to 0.45%, respectively.[4] CFE occurs in the aortic valve in 29%, the mitral valve in 25%, the tricuspid valve in 17%, and the pulmonary valve in 13%.[5]

Patients are mostly asymptomatic. Although it is histologically benign, it varies according to the location, size, growth rate and embolization tendency of the tumor; It can cause life-threatening complications such as stroke, valve dysfunction, embolism, myocardial infarction, pulmonary embolism and sudden death. Mechanisms of thromboembolism include tumor embolization and thrombus formation on the tumor. Tumor size may not be an indicator for thromboembolism risk.[6]

Warfarin or antiplatelet therapy can be used in the treatment of small and asymptomatic lesions and may be useful in preventing thromboembolic events.[7] However, surgical removal of tumors larger than 1 cm is recommended due to the risk of embolization and sudden death.[4] Although the need for surgery in small and asymptomatic cases is controversial, embolization has also been reported in a 3 mm tumor.[8] In our case, the size of the tumor was more than 1 cm and we excised it surgically because it was symptomatic. After the first operation, valve functions were observed to be normal and the patient was discharged without complications. However, two years later, recurrence developed in the same place. This time, aortic valve functions were also observed to be impaired. For this reason, this time we had to remove the aortic valve along with the mass, and therefore we performed AVR.

Recurrence is not common because CFE is benign. In our case, the first complaint of the patient was distal arterial embolism, which developed at close intervals and twice. At his second visit, in addition to the recurrence of the tumor, ischemic findings and infarct areas were also detected in the kidney and spleen. Since the coexistence of these conditions is very rare, we think that this case report contributes to the literature. Therefore, cardiac evaluation is extremely important in patients presenting with embolism in the extremity arteries and visceral organs. In the case of recurrent and multiple embolism in patients with a previous history of cardiac tumors, it should be kept in mind that benign tumors may also develop recurrence, and imaging methods such as MR or transesophageal echocardiography (TEE) should be investigated. In addition, the surgical material should

be examined pathologically in patients presenting with embolism.

CONCLUSION

Although cardiac fibroelastomas are benign, recurrence can be seen. It can cause embolization and should be kept in mind in the differential diagnosis of patients presenting with embolism. Although there is no definite consensus on surgical removal in asymptomatic patients, they should be surgically removed in symptomatic patients.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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